Brugada Syndrome

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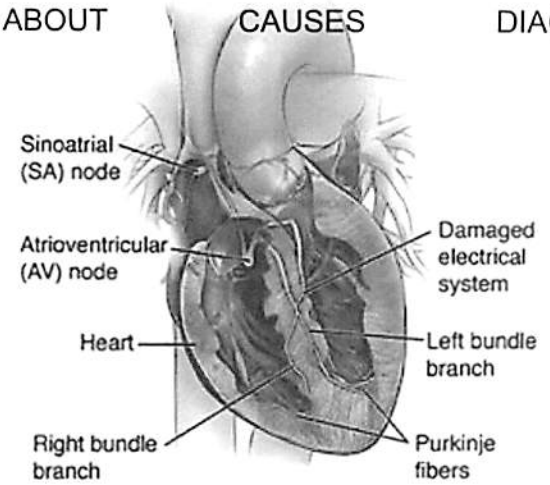
What is Brugada syndrome?

Brugada syndrome is a genetic disorder that can cause a dangerous irregular heartbeat. When this happens, the lower chambers of your heart (ventricles) beat fast and irregularly. This prevents blood from circulating correctly in your body. This can be dangerous and may lead to fainting or even death, especially during sleep or rest. The disease has been called sudden, unexplained nocturnal death syndrome because people with it can often die in their sleep.

Damaged electrical system of the heart



Brugada syndrome is rare. It affects about 5 of every 10,000 people worldwide. Symptoms often start during adulthood. But the disorder can develop at any age, including infancy. The average age of death related to the disease is 40 years old.

What causes Brugada syndrome?

The genetic form of Brugada syndrome is most often caused by a defect in the SCN genes. But other genes can be involved, too. It can be inherited from just one parent. But some people develop a new defect of the gene rather than inheriting it from a parent. These genetic defects cause malfunctioning sodium channels in the heart muscle cells. This may lead to abnormal heart rhythms.

It's possible to have Brugada syndrome that stays inactive (dormant) and doesn't cause any problems. But some medicines such as antidepressants and antipsychotics, illegal drugs, conditions that cause fever, and electrolyte problems can unmask the syndrome. Some people may seem to have Brugada syndrome based on certain characteristics seen on an electrocardiogram. But they don't actually have the disease itself. This is called a Brugada ECG pattern. It may not pose a risk if the condition is short-term (temporary) and doesn't cause symptoms or dangerous heart rhythms.

Who is at risk for Brugada syndrome?

People who are at greatest risk for the disorder are those of Asian descent, particularly Japanese and Southeast Asian heritage. It occurs 8 to 10 times more often in men than in women. Researchers think the male hormone testosterone may contribute to the difference between genders.

What are the symptoms of Brugada syndrome?

Symptoms that may occur with Brugada syndrome include:

Palpitations

Fainting

Sudden death or cardiac arrest

How is Brugada syndrome diagnosed?

Your healthcare provider may think you have Brugada syndrome if you have any of the above symptoms not explained by another more common condition. They may also suspect it if you have a family history of the disease, or an unexplained sudden death in the family. An electrocardiogram (ECG) can help diagnose the disease. People with typical ECG features are often diagnosed with this condition if they have had:

Fainting

Sudden cardiac death

Fast heart rate starting from the heart's bottom chambers (ventricular tachyarrhythmia)

Family history of sudden cardiac death or Brugada syndrome

Genetic testing can also be done to help diagnose the condition.

If the diagnosis is unclear or if you are found to have the Brugada ECG pattern but have no symptoms, you may have a test called a "drug challenge." This is done in a hospital setting with professional supervision. You are given a medicine that blocks sodium channels. Then several ECGs are done to look for changes in your ECG.

First-degree relatives of someone with Brugada syndrome should be screened. This may include at least a health history, physical exam, and an ECG. Genetic screening may also be used.

How is Brugada syndrome treated?

Currently, there is no cure for Brugada syndrome. But there are ways to protect people from the dangerous consequences of the disease. An implanted cardioverter defibrillator (ICD) can help prevent sudden death linked to Brugada syndrome. When this device detects the start of an arrhythmia, it will either try to stop it with pacing or deliver a shock to reset it back into its regular rhythm.

Medicines may also help prevent arrhythmias. Another possible treatment option may be a cardiac ablation. In this procedure, a small area of heart tissue found to cause the dangerous arrhythmia is destroyed. Discuss all options with a qualified healthcare provider.

It's also important to stay away from certain medicines that can make Brugada syndrome worse.

What are possible complications of Brugada syndrome?

The most serious complication of Brugada syndrome is sudden death. This often occurs while the person is sleeping.

What can I do to prevent Brugada syndrome?

Many cases of Brugada syndrome are related to a genetic defect. It's not possible for you to prevent inheriting this condition. But identifying the condition is key to preventing its possible complications. If you have Brugada syndrome and plan to have children, you may want to consult with a genetic counselor first.

When should I call my healthcare provider?

If you have any of the symptoms of ventricular arrhythmia, get emergency medical help right away. If you think you may be at risk for Brugada syndrome because of your family history or other reasons, see a healthcare provider for testing.

Key points for Brugada syndrome

- Brugada syndrome is a genetic disorder that causes an irregular heartbeat.
- It can be either inherited or acquired.
- Genetic testing can look for a mutation such as with the SCN5A gene that may help determine your risk.
- If Brugada syndrome is diagnosed, an implanted cardioverter defibrillator (ICD) may be considered to prevent sudden death linked to the syndrome.
- If you have any of the symptoms of ventricular arrhythmia, get emergency medical help right away.

Next steps

Tips to help you get the most from a visit to your healthcare provider:

- Know the reason for your visit and what you want to happen.
- Before your visit, write down questions you want answered.
- Bring someone with you to help you ask questions and remember what your provider tells you.

At the visit, write down the name of a new diagnosis, and any new medicines, treatments, or tests. Also write down any new instructions your provider gives you.

Know why a new medicine or treatment is prescribed, and how it will help you. Also know what the side effects are.

Ask if your condition can be treated in other ways.

Know why a test or procedure is recommended and what the results could mean.

Know what to expect if you do not take the medicine or have the test or procedure.

If you have a follow-up appointment, write down the date, time, and purpose for that visit.

Know how you can contact your provider if you have questions.

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